Diagnosis of Disorders of the Cerebellar Apparatus

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From
THE ARCHIVES OF DIAGNOSIS
January, 1910.

Extracted from "The Archives of Diagnosis," New York, January, 1910

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Sherrington[1] has given the name proprioceptive system to that part of the nervous apparatus which is specialized to receive impressions as to the position and condition of the body itself irrespective of external stimuli.

Anatomy.—The system of nerves of which it is composed arises in end-organs situated in tendon, aponeurosis, muscle, and perhaps cartilage, bone and subcutaneous tissue, as well as possibly in the parietes of the body cavities, although this is not yet definitively known. At all events, the impressions are conveyed in fibers most of which accompany the motor nerves until they leave them to enter the intervertebral ganglia on the posterior roots of the spinal cord. The fibers then enter the spinal cord via the posterior roots. Their course from this point is only in part ascertained. It is certain that some of the impulses eventually pass upwards homo- and contralaterally in the dorsal and ventral direct cerebellar tracts respectively, having first passed through a relay neurone, probably in the column of Clark. Some observers believe that other impulses of the proprioceptive system run in the posterior columns as far as the nuclei of these in the medulla oblongata, and that from there, the impulses proceed in the arciform decussation until they partly in turn reach the cerebellum, like the direct tract, in the restiform body, and partly, along with the ventral cerebellar, accompany Gowers' tract and then that some pass through the brachium conjunctivum, others proceeding by the brachium pontis.

But it is difficult to determine this, for it is quite certain that the main part of the impulses passing along the dorsal columns are those which subserve the conscious sense of attitude, while in the main the spino-cerebellar impulses do not directly affect consciousness, although Van Gehuchten[2] believes that even the common sensory tract passes in part via the cerebellum, to eventually reach the

cortex through a cerebello-fugal path which decussates in the brachium conjunctivum en route to the opposite cortex. This view, however, is not generally accepted.

The impulses which reach the cerebellum in these various ways are there coordinated; and as a result of their impression, efferent impulses pass from the Purkinje cells on their way to regulate body tonus and position in accordance with the information gained.

The efferent tract first reaches the dentate body; and it from there intromits via the brachium anterius with a second relay neurone in the red nucleus of the opposite side, whence it again decussates in Forell's bundle and passes through the tegmental region of the mesencephalon, becoming more ventral and lateral in the pons, bulb and cord, in the latter more or less commingling with the anterio-lateral portion of the lateral pyramidal motor tract, and is supposed, with it, to mediate the activity of the motor cells in the anterior cornua of the spinal cord.

It is supposed that other cerebello-fugal fibers control the ponto-bulbar nuclei in a similar manner, and thus regulate the activities of the muscles supplied by the motor cranial nerves. But the information by which the position of head and eyes is mainly regulated has another organ much more sensitive than the end-bodies in the extremities, neck and trunk. This is situated in the membranous labyrinth and consists of the terminations of the vestibular nerve there. Impressions are communicated to this by the movements of the endolymph within the utricle and semicircular canals. Thence they pass via the vestibular nerve to the nucleus of that name in the medulla, and from there probably in the lateral lemniscus and brachium conjunctivum to the homolateral and contralateral cerebellar lobe and vermis; some of them, however, pass to the opposite oculomotor nuclei, and others directly to the nuclei of Deiters and von Bechterew.

Physiology and Pathology.[3]—The receptive function of the cerebellum is firstly orientation of the different parts of the body with regard to one another, and secondly perhaps a participation in the orientation of the body to its surroundings. It is possible, however, that this latter function is entirely cerebrospinal and that the importance of the cerebellum in relation to it is confined to its proprioceptive aspect.

What Sherrington calls the ejicient function of the cerebellum is that in virtue of which it coordinates and harmonizes body, head and eye movements by means of its efferent fibers. It is in this way that the equilibrium of the body is automatically regulated.

Thus, the disorders of the cerebellar apparatus lead to disorientation and disequilibrium. And it is the disorder of these together which constitutes vertigo. This is defined by Grasset[4] as "a subjective psychical phenomenon constituted by the transmission to the center O* of a double basilar sensation (mesencephalic and cerebellar), a false sensation coming from the apparatus of orientation, and a sensation of insufficience of the automatic centers to assure the equilibrium." Pierre Bonnier,[10] on the other hand, separates the two elements thus united by Grasset, and distinguishes vertigo from a vertiginous sensation, while Hughlings Jackson[11] gives the name of vertigo only to the motor element, that is the beginning of the fall.

However, intellectual assent to the illusion of displacement of objects or autorotation cannot be regarded with Pierre Bonnier as essential. The real essential is not an intellectual belief in one's disorientation, but a sentiment of loss of equilibrium; for this may be felt in bed when the patient knows perfectly that he is not falling. Thus true vertigo cannot occur except from disorder of the cerebellar apparatus, which is the organ of both the functions the perturbation of which constitutes vertigo.

In addition to these essentials, accessory and inconstant phenomena may accompany vertigo. Some of these are due, following Bonnier, to the overflow of highly charged nerve centers when powerfully stimulated, their ensemble constituting the bulbar syndrome[12] which he described. For instance, when the vestibular nerve is powerfully irritated, as by extension of otitis media to the

*By the symbol O, Grasset[5] represents what he calls the superior psychical center or centers in which consciousness is achieved. He distinguishes these from other cortical centers which only subserve sensibility and psychomotricity and even coordinate action and ideation when these are only automatic or subconscious and which he designates by the symbol ⬡, the *polygon*. He believes that this schema represents a real hierarchy of the nervous centers. It is a restatement of the "high and low-level" hypothesis of Hughlings Jackson.[6] Recent studies of apraxia have added concreteness to this conception; but the phenomena of hysteria and so-called psychical

labyrinth, the turning sensation and loss of equilibrum may be accompanied by nystagmus or even conjugate deviation from stimulation of the oculo-motor nuclei which are intimately connected with the vestibular nucleus by means of the posterior longitudinal bundle. If the stimulus extends to the pneumogastric nucleus, nausea, vomiting, palpitations, dyspnea and even syncope may occur. Noises in the ears are very common, even when the irritation does not originate auricularly. These bulbar phenomena are very often characterised by the state of angoisse, with acute distress, horror and fear of impending death such as to resemble angina pectoris.

The source of vertigo is detected clinically by its aggravation through putting into action of the organ at fault, and its cessation or improvement when that organ is at rest: thus labyrinthine vertigo diminishes when the patient is quiet in bed, ocular vertigo when the eyes are closed, vertigo of gastric origin when the stomach is at rest, cardiac vertigo when strain on the heart is diminished, and so on; while vertigo of central origin is not attenuated by any of these means in particular.

The seat of this disorder need not be in the cerebellum itself; a lesion in any of its peduncles is as effective: so that in the case of intracranial neoplasm, vertigo is a localizing sign only in a very broad sense. Thus the vertigo of insular sclerosis is usually due to patches situated in the pons or other brachium, rather than to a lesion in the cerebellum itself.

The vertigo occasionally found in tabetics is really a peripheral one due to the implication of the vestibular nerve in the meningeal process which causes tabes of the posterior columns by implicating the posterior roots. When the labyrinthine impulses are thus interfered with, the test with voltaic electricity is applicable. This is performed by placing one pole of a galvanic battery over each mastoid process. In the normal individual, the head deviates, as soon as the current is closed, towards the positive pole; while there is felt

disaggregations upon which Grasset largely based his schema can no longer be considered as supporters of his hypothesis, now that the work of Babinski,[7] Sidis[8] and Bernheim[9] has shown the fallaciousness of the data by which they were explained; so that Grasset's schema, like those concerning the centers for speech, is dangerous to the uninitiated in that it tends to make one force the facts to fit the fancy.

a sensation of surrounding objects being displaced towards the positive pole. If either labyrinthine apparatus or center is interfered with this phenomenon does not occur when the positive pole is placed on the diseased side. These sensations and movements are accompanied by nystagmus towards the positive pole.

Other means of exploring the vestibular function are the irrigation test of Barany[13] and the rotation platform, in which nystagmoid movements occur long before vertigo is produced. When a person is seated on a platform revolving towards the right, the eyes will be observed, or better felt, to make a series of rapid rhythmical movements in the same direction as that in which he is turning, after each jerk returning more slowly to the mid position. After a few seconds, these movements cease until the platform ceases to turn, when they recommence, but in the opposite direction. These movements are dependent upon the inertia of the endolymph, as was shown experimentally by Ewald.[14] Their non-occurrence signifies interference with vestibular function, provided of course the oculo-motor apparatus is intact. The right labyrinth is the main controller of rotation to the right and vice versa, as the following case shows:

After an accident, a workman felt out of sorts, but had only slight pain on side of head; but on questioning, he admitted sounds in the ear and vertigo when upright, objects seeming to turn from left to right. On testing, there was very little nystagmus in rotation to the right, but much more after rotation to the left. The confirmation of the lesion of the left vestibule was gained by a demonstration of diminished perception of sound in the left ear, both to aerial and cranial conduction[15] (Goldman of Cairo).

The irrigation method does not depend upon pressure, but on temperature. Cold water causes nystagmus towards the opposite side, but the test is less responsive than that with hot water at 40° C. unless the tympanum is perforated. These effects are explained by the induction of currents in the endolymph by the change of temperature. Thus, cold water causes a current from the top of the anterior semicircular canal to the ampulla below; and hence when the right ear is injected, a rotation nystagmus to the left is produced. It is only when the head is leant on the shoulder so as to tilt the horizontal canal into a more vertical plane that this test causes lateral nystagmus: because in the horizontal plane gravity does not displace

endolymph even when its density is changed by our test. Thus the test is in reality a mechanical, and not a thermal one. There is a strong probability that some cases of drowning are due to the vertigo produced by stimulation of the labyrinth through the cold water entering the ear.

Tests and Diagnosis.—As may be readily imagined, the disorders of so extensive and complex a function should be frequent, and should manifest themselves clinically. They may be summed up in the term physical disorientation total or partial, global or focal; and the most familiar example is the staggering walk of intoxication. Less frequent, though more dramatic, is the sudden fall of Menière's disease, in which the patient, suddenly deprived of knowledge as to the position of the head, is utterly lost, and hence falls in a heap. Both these examples signify interference with the afferent cerebellar function.

When the efferent[16] function is interfered with, muscular tonus is completely lost, and the deep reflexes disappear. Moreover, there develops an incapacity to rapidly alternate movement—adiadocokinesis; and the synergia of different muscles is much impaired; so that a coordinate movement of two or more joints is only performed slowly and jerkily—coarse intentional tremor. At rest, on the contrary, the power of immobilization is much increased. Although there is no actual weakness, yet a virtual asthenia arises from the atonic state of the muscles. The effort needed to overcome this produces rapid fatigue.

The result of the asynergia is the clumsy infantine walk which the patient adopts for safety and the extensive shaking of the hand when any movement is attempted, often preventing writing or even the use of a knife and fork. The same asynergia, affecting the tongue, palate and facial muscles, produces slow, monotonous, toneless and sometimes palatal and explosive speech. It is shown in the eye muscles by nystagmus. If dysergia is only slight it may be elicited by—

a. Rapid pronation and supination of the hand being more slow or clumsy on the affected side.

b. By failure to bend the knees when the patient from the standing position is made to lean far back.

c. By a jerky and needlessly high lifting of the knee when the patient endeavors to kneel upon a chair.

d. By a tendency of the thigh to flex when the patient sits up from recumbency.

e. By the usual finger to nose test, which often reveals the tendency to pass the point aimed at, the hand sometimes violently knocking the face.

f. When the patient, in drawing a straight line, tries to arrest the movement at a fixed point, he will be unable to do so, the pencil traveling beyond. I have devised a practical application of this test as follows: A horizontal line is drawn across a piece of paper. A perpendicular is let down from its left extremity. Three vertical marks are made at equal distances along the horizontal line. The patient is then directed to draw, beginning at the vertical line, three horizontal lines, one below the other, and each stopping abruptly at the 1st, 2d or 3d vertical mark, respectively. They must be drawn rapidly, and each with a single movement. The normal person will pass the limit only very slightly, and will make hardly any returning stroke or movement of the arm. Cerebellar dysergia is indicated by excessive length, and especially by a too great movement of recovery whether the paper is marked or not.

Increased power of stabilization often characterizes cerebellar cases. It is ascertained by placing the patient on his back with his legs raised in the air and noting the ease and duration of maintenance of an attitude difficult and fatiguing for normal individuals, in whom, moreover, there is always a certain amount of rocking to and fro, whereas the cerebellar case appears motionless.

This test is a very valuable one against the ataxia of tabes dorsalis, in which condition the tendency to oscillate from the stationary position is much exaggerated.

From ataxia in general, dysergia of cerebellar type can moreover be differentiated by the effect of vision upon the patient's movements. True ataxia is much accentuated when the patient closes his eyes, which procedure does not influence dysergia due to interruption of efferent cerebellar impulses. Again, a tabetic, when he will, can arrest any movement at any point desired, provided that he receives knowledge of the whereabouts of his limb by another sense than that of attitudes, which in him is impaired. It is upon this principle, in-

deed, that depends the commencement of the Fraenkel method of re-education by graduated movements. The cerebellar, on the other hand, does not appear to be reeducable, to judge from the few chronic cases which have been carefully enough observed.

Another character of tabetic ataxia is its irregularity, which distinguishes it from the mere unmeasuredness of the movements of the cerebellar patient, in whom the walk is like that of the drunkard.

Another valuable distinction is that in the standing position the cerebellar case merely oscillates as if balancing, the feet remaining tranquilly on the ground and there being none of the violent muscular contractions which are seen in the legs of a tabetic when endeavoring to stand. It is thus entirely erroneous to give the name ataxia to the results of a defect of the cerebello-fugal system.

The combination of an exaggerated static equilibrium, sometimes cataleptoid in degree, with dysergia leading to intentional tremor produces a singular clinical picture, in some respects resembling that of paralysis agitans with intention tremor.

The distinction of the cerebellar syndrome from ataxia is no mere academic one, but is of enormous clinical importance in certain affections, as it enables the physician to diagnose the site of a lesion which he might otherwise fail to locate.

But of course the cerebellar syndrome in its purity does not indicate the level at which the cerebellar apparatus is implicated: it does not inform us whether the fault is in the cortex of the cerebellum, the radiations in the white matter, Deiter's nucleus, the brachium conjunctivum, the nucleus ruber, or in the rubrospinal tract whether in mesencephalon, pons, medulla or spinal cord. The method of finding this out is that employed in locating lesions in the psychomotor, sensory or any other physiological tract-system of the cerebrospinal axis. It is a topographical method, consisting of a clinical study of physiological systems contiguous to the one manifesting symptoms. Thus in the cord, when extra cerebellar, the rubrospinal tract can hardly be affected without implicating the pyramidal fibres, and thus producing spasticity, hyperactive reflexes, combined flexion or other homolateral anergias, and most important of all the hallux extension when the sole is stroked.

Moreover, the Gower's column may be implicated and give rise to contralateral sensory dissociation of the syringomyelic type, the

cutaneous position of which will indicate the cord level of the lesion, which is at a point about seven segments higher up than the uppermost segment of the cutaneous loss.

When the lesion is in the bulb, the roots or nuclei of the 12th, 11th, 10th and 9th cranial nerves are hardly likely to escape; and one may look for atrophy of the tongue, respiratory symptoms or insufficiency of the trapezins, sternomastoid or laryngeal muscles. Higher up still, towards the pons, anomalies of the facial and abducens innervation may guide us, or the vestibular function may be affected at its relay neurone here. Still higher, in the mesencephalon, the pathetic and oculomotor nerves may become involved at their origin, and the optic fibers themselves may be implicated near the colliculous by the lesion. In any part of its long course, the central origin of the 5th nerve may participate; and in a similar way the lemniscus may be shown by sensory loss, and the psychomotor tract by perturbations of the motility to be symptomatically involved by the transverse extension of a lesion of the rubrospinal tract.

When no localizing symptom of this kind is found, marked dysergic symptoms are very apt to be localized in the cerebellum itself. Now the cerebellum is a homolateral organ, and hence the symptoms may be entirely onesided, a great help in diagnosis; but even in this situation, we have means of localization more certain than exclusion; for a lesion within the cerebellum large enough to produce dysergia is generally large enough to affect the intracranial circulation either directly or by causing an increase of subtentorial pressure. This causes ventricular dilatation, and gives rise to the well-known picture so conspicuous a feature of intracranial neoplasm. It is constituted clinically by headache, nausea, projectile vomiting, etc.; but the consideration of these features is another chapter of neural semeiology.

The Diseases which produce dysergia.—I conclude with a short statement of the conditions in which disorder of the cerebello-fugal apparatus is most frequent.

Disseminated sclerosis. The alert reader will have been struck by the resemblance of the clinical picture to that of insular sclerosis, of which the slow, explosive speech, the nystagmus and the intentional tremor are indeed indices of the incidence of the multiple sclerosis upon the rubrospinal tracts.

Paralysis agitans. The tremor, slow and toneless speech, fixed facies and antero latero- or retro-pulsion of Parkinson's disease are all symptoms referable to the cerebellar apparatus. And the lesions of the brain stem shown by Spiller and Camp[17] may well account for this.

Paresis. Frequently the walk of the general paralytic has a cerebellar character, with its wide-apart feet, festooning pattern and uncertain tendency. It is explained pathologically by recent researches,[18] which have shown that the cerebellar cortex invariably takes its place in the general meningo-encephalitis of this disease.

Tumors. To neoplasms I have already alluded; and analogous remarks might be extended to intracranial abscess.

Hereditary maldevelopments. Finally, the congenital agenetic affections must be borne in mind; for though cases have been reported which came to autopsy with marked cerebellar atrophy which had never produced the least clinical sign, yet, there is a group of familiar dystrophies allied to Friedreich's ataxia in which the cerebellum is conspicuously abnormal and which does exhibit symptoms in life similar to those described in the preceding pages.

Toxemia. Finally, intoxications may produce a syndrome made more complex by perturbations of the cerebellar apparatus.

Thus, the cerebellar syndrome, clear cut though it be, is no exception to the common complexity and difficulty of the diagnosis of disease. Its knowledge gives us another arm, to learn the use of which still more study and skill are exacted of the physician of our day.

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